



**University of
Zurich**^{UZH}

**Zurich Open Repository and
Archive**

University of Zurich
University Library
Strickhofstrasse 39
CH-8057 Zurich
www.zora.uzh.ch

Year: 2015

Is the desire for amputation related to disturbed emotion processing? A multiple case study analysis in BIID

Bottini, G ; Brugger, P ; Sedda, A

Abstract: Body integrity identity disorder (BIID) is characterized by the overwhelming desire to amputate one or more healthy limbs or to be paraplegic. Recently, a neurological explanation of this condition has been proposed, in part on the basis of findings that the insular cortex might present structural anomalies in these individuals. While these studies focused on body representation, much less is known about emotional processing. Importantly, emotional impairments have been found in psychiatric disorders, and a psychiatric etiology is still a valid alternative to purely neurological accounts of BIID. In this study, we explored, by means of a computerized experiment, facial emotion recognition and emotional responses to disgusting images in seven individuals with BIID, taking into account their clinical features and investigating in detail disgust processing, strongly linked to insular functioning. We demonstrate that BIID is not characterized by a general emotional impairment; rather, there is a selectively reduced disgust response to violations of the body envelope. Taken together, our results support the need to explore this condition under an interdisciplinary perspective, taking into account also emotional connotations and the social modulation of body representation.

DOI: <https://doi.org/10.1080/13554794.2014.902969>

Posted at the Zurich Open Repository and Archive, University of Zurich

ZORA URL: <https://doi.org/10.5167/uzh-101083>

Journal Article

Accepted Version

Originally published at:

Bottini, G; Brugger, P; Sedda, A (2015). Is the desire for amputation related to disturbed emotion processing? A multiple case study analysis in BIID. *Neurocase*, 21(3):394-402.

DOI: <https://doi.org/10.1080/13554794.2014.902969>

Running title: BIID and emotion

Title: Is the desire for amputation related to disturbed emotion processing? A multiple case study analysis in BIID.

Gabriella Bottini^{1,2}, Peter Brugger³ & Anna Sedda^{1,2}

(1) Behavioural and Brain Science Department, University of Pavia, Italy

(2) Cognitive Neuropsychology Laboratory, Niguarda Ca' Granda Hospital, Milan, Italy

(3) Department of Neurology, University Hospital Zurich, Swiss

Corresponding Author:

Gabriella Bottini;

DBBS - Piazza Botta 11, 27100 Pavia, Italy

Email: g.bottini@unipv.it.

Acknowledgement

The authors thank Prof. Anne Schienle (Graz, Austria) for providing the German version of the Disgust Scale. We are indebted to two anonymous referees for their highly constructive critique.

Funding

This work was supported by QUALIA – Associazione per la ricerca teorica e applicata in scienze cognitive, to [A.S.].

Disclosure of Conflicts of Interest

None of the authors has any conflict of interest to disclose. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this report is consistent with those guidelines.

Abstract

Body Identity Integrity Disorder (BIID) is characterized by the overwhelming desire to amputate one or more healthy limbs **or to be paraplegic**. Recently, a neurological explanation of this condition has been proposed, in part on the basis of findings that the insular cortex might present structural anomalies in these individuals. While these studies focused on body representation, much less is known about emotional processing. Importantly, emotional impairments have been found in psychiatric disorders, and a psychiatric aetiology is still a valid alternative to purely neurological accounts of BIID. In this study, we explored, **by means of a computerized experiment**, facial emotion recognition and emotional responses to disgusting images in seven individuals with BIID, taking into account their clinical features and investigating in detail disgust processing, strongly linked to insular functioning. We demonstrate that BIID is not characterized by a general emotional impairment; **rather, there is a selectively reduced disgust response to violations of the body envelope**. Taken together, our results support the need to explore this condition under an interdisciplinary perspective, taking into account also emotional connotations and the social modulation of **body representation**.

Keywords: Body Integrity Identity Disorder (BIID), Apotemnophilia, Xenomelia, Somatoparaphrenia, Embodied emotions.

Introduction

Persons with Body Identity Integrity Disorder (BIID) feel a desperate urge to have one or more healthy limb amputated (First & Fisher, 2012) or, as recently highlighted, to become paraplegic (Giummarra et al. 2012). This disorder was initially known as “apotemnophilia”, “love for amputation”, and considered part of the larger category of paraphilias (First & Fisher, 2012). However, the desire for amputation in BIID is primarily driven by the need to have a more “complete” body shape and only in some cases by a sexually motivated compulsion (First & Fisher, 2012). Psychiatric, psychodynamic and psychological hypotheses have been put forward to explain BIID (First & Fisher, 2012). However, the condition remains elusive and difficult to be defined respecting all its heterogeneous features. For these reasons, BIID is not included in the DSM-IV, in contrast to Body Dysmorphic Disorders (BDD), which is listed as a “hypochondriac disturbance”. BIID and BDD subjects only share the overwhelming frustration associated with their own bodily appearance. However, persons with BIID are perfectly aware that the point of concern about their own bodily appearance is a purely subjective issue. On the other hand, persons with BDD are preoccupied with particulars of their appearance, which they normally think are also available to others (Phillips et al 2005). Another distinctive feature between the two disorders (Ryan & Shaw, 2011) is that individuals with BIID are never ashamed of their limb, even though this disturbance seems to have the same pervasive social impact as BDD, frequently causing a chronic dysphoria and a persistent sense that their “able-bodied status” is inappropriate (First & Fisher, 2012). The resulting urge to get rid of their undesired limbs is so intolerable to make persons with BIID looking for complaisant surgeons or even causing an accident involving the non-accepted limb (Bou Khalil & Richa, 2012).

Only recently BIID has been interpreted as a neurological impairment (Brang, McGeoch, & Ramachandran, 2008), due to some similarities of this condition with other disorders of body representation (Sedda, 2011). Neuroimaging studies on individuals with BIID identified structural and functional impairments of the parietal lobe and of the insula, both considered crucial areas for body representation (Hilti et al., 2013; McGeoch et al., 2011). Followers of this neurological hypothesis propose to move from BIID to “xenomelia”, a Greek word meaning “foreign” and “limb”, to parallel somatoparaphrenia (McGeoch et al., 2011). For both these disorders, a common anatomical pattern has been found, involving the right parietal cortex and the right insula (Gandola et al., 2012; Giummarra, Bradshaw, Nicholls, Hilti, & Brugger, 2011). Somatoparaphrenia is a delusion confined to the contralesional paralyzed limb, causing an intense feeling of non-belonging and the ascription of the own arm or leg to family members or caregivers (Gandola et al., 2012). However, somatoparaphrenic patients do not typically express a desire for limb amputation (Bottini et al., 2009), and the similarities between the disorders may be rather superficial (Sedda, 2011). Moreover, methods to transiently abolish somatoparaphrenia do not help reintegrate the affected limb in BIID (Lenggenhager et al., in press).

The puzzling attitude of persons with BIID to amputation or plegia may well involve an emotional component, in particular the feeling of disgust for body violations (Rozin, Haidt, & Fincher, 2009). Although classically considered a primary emotion of protection from potentially harming or contaminating food, disgust has been reinterpreted as a multimodal construct (Rozin et al., 2009), so that poor hygiene, inappropriate sexual acts, death and, finally, violations of the ideal body “envelope” or exterior form (Rozin et al., 2009) are considered additional domains. Interestingly enough, bodily cues that cause inferential thinking about disease and characterize an individual as a possible disease carrier have been shown to induce an automatic reaction of disgust. For instance, obesity elicits strong aversion in people with concerns about pathologies, and some of the obsessive

compulsive disorder variants (i.e. contamination based) are strongly connoted by a dysfunction of the disgust system (Curtis, 2011).

The insula appears a crucial component of the body representation network, particularly for body awareness (Berlucchi & Aglioti, 2010), and of disgust processing (Jabbi, Bastiaansen, & Keysers, 2008).

Individuals with BIID might not consider the amputation of one's own limb as an unacceptable violation of the ideal body "envelope". Furthermore, they are deeply convinced that the severe physical disability following the amputation would represent their ideal state of being (First & Fisher, 2012). As a consequence, they might not experience disgust for the amputation of this limb. To our knowledge, this putative emotional component and the integrity of the disgust system has never been investigated in BIID.

The actual debate on BIID hypothesizes also that this disturbance might be induced by a psychiatric disorder with a wider emotional impairment (De Preester, 2011). Consequently, individuals with BIID might manifest a general pathological emotional processing. If this is the case, not only body violations, but also domains related to the contamination core (blood, hygiene) should be compromised. On the other hand, if BIID derives from a derangement selectively concerning body representation, they should present with a specific/isolated emotional impairment. This deficit should be related to this cognitive aspect despite an otherwise intact emotional processing.

In this study, we investigated facial emotion recognition and emotional responses to disgusting images in a group of individuals with BIID to test the hypothesis of a selective impairment of the emotional component for stimuli related to the body envelope and not for stimuli concerning other domains. Considering our experimental tasks, the first hypothesis suggests that if BIID is induced by a psychiatric disorder, subjects should be generally less accurate in recognizing facial emotions (Kohler, Hoffman, Eastman, Healey, & Moberg, 2011), should rate images depicting disgust related to the contamination core as less disgusting than controls (Curtis, 2011) and should be more disgust

sensitive than the average population (Schienle et al. 2013). In contrast, if BIID selectively compromises body related stimuli only, one should expect a regular accuracy in the facial emotion recognition task, a normal rating for non-body stimuli (independently from the category) and a disgust sensitivity comparable to that of controls. In parallel, this hypothesis predicts diminished disgust to violations of the ideal body envelope, given that individuals with BIID experience that their actual body envelope is far from ideal. In fact, one could anticipate such a result given that the ideal body form in a person with BIID is so far apart from those cultural, social and biological norms that usually trigger a disgust response when violated (Rozin et al. 2009).

Methods

Patients

Seven self-claiming individuals with BIID were administered with a facial emotion recognition (FER) task, a picture rating task (PRT), the Disgust Scale (DS) and a semi-structured psychiatric interview (Table 1).

Six subjects were men. All male subjects manifested BIID for their legs, while the female subject wanted to have her left arm removed. Two subjects were from Switzerland, one was Italian and four were German. Individual consent was obtained according to the Declaration of Helsinki (BMJ 1991; 302: 1194) and the University of Pavia had approved the study. Subject 4 was left handed, while all the other subjects were right handed.

[Insert Table 1 here]

Control Group

Control participants were healthy subjects recruited at the University of Pavia, who previously participated in either the tasks to obtain a normative group, independently from this study. Subjects were referred on the basis of absence of previous history of mental or neurological illness,

sensorimotor impairments, mood disorders and drug treatment. All subjects were right-handed and with normal or corrected-to-normal vision. Individual consent was obtained according to the Declaration of Helsinki (BMJ 1991; 302: 1194) and the University of Pavia approved the study.

From a larger dataset ($n=20$ for both tasks), subjects **were identified** based on education, age and gender, to obtain a control sample matched to individuals with BIID. The selection based on demographic features led to a different sample size for the two tasks. The control group for the FER task included 10 males (mean age: 29.9 ± 8.08 ; mean education 14.3 ± 3.9) and 8 females (mean age: 32.75 ± 8.22 ; mean education 14.8 ± 2.58). Seven males (mean age: 32.4 ± 4.99 ; mean education 15.1 ± 2.67) and 7 females (mean age: 36 ± 9.96 ; mean education 16.2 ± 3.25) composed the control group for the **PRT**. Statistical comparisons confirmed that both control groups are not significantly different compared to individuals with BIID in age and education (all $p > 0.05$).

Materials & Procedure

Semi-structured psychiatric interview

The initial interview was modelled after the SCID-II preliminary questionnaire (First, Gibbon, Spitzer, Williams, & Benjamin, 1997). We complemented our interview with questions concerning family (i.e. whether parents and relatives know about the disease), psychotherapy treatment, mood, compulsions and obsessions concerning the desire or other domains, and sexual orientation.

FER task

Images were taken from the Ekman and Friesen series (Ekman & Friesen, 1976). The task included 3 displays for each emotional category (fear, sadness, disgust, happiness and anger), for each individual (JJ and MO, two individuals, one male and one female) (Figure 1A Supplementary materials). Facial emotions, **obtained using a morphing technique**, were depicted at 25% and 50%

of intensity, to detect even subtle emotional recognition deficits (Sedda et al., 2013), leading to a total of 60 trials for this task. We excluded “surprise” from the emotion set as previous studies reported that even healthy subjects frequently confound this emotion with fear (Rapcsak et al., 2000).

We presented the FER task on a 13.3-in. computer screen (Asus Zenbook, Windows) with a screen resolution of 1024×768 pixels. Stimulus presentation was controlled through a custom developed Matlab© script. On each trial, one image was shown in the centre of the screen (randomized order). With their dominant hand, participants had to press the keyboard label that corresponded to the emotion depicted on the image, as quickly and accurately as possible. Labels were placed on the computer keyboard in a randomized order across participants, from key "t" to key "o" of an Italian keyboard, each label depicting the initial of the verbal label for the emotion (participant's mother tongue). Before the experiment, subjects practiced to recognize the corresponding initials (e.g., A for anger, F for fear). Starting point for the responding hand was always in the same position, indicated by a cross placed on the computer touchpad. Subjects were instructed to return their hand to the starting position between trials. Between trials, a fixation cross with a variable inter-stimulus interval appeared at the centre of the screen. There was no time limit to complete a trial. For each trial, accuracy was collected. The experiment took about 20 minutes to complete.

PRT

This task was composed of pictures depicting fear, sadness, happiness and anger scenes as control stimuli, and by pictures displaying disgust (Figure 1B Supplementary materials). This experiment has been modeled following Rozin model on disgust (Rozin et al., 2009) and similar tasks employed in other experiments exploring the disgust system (Schienle, et al., 2013). Participants are presented with 36 real-life images, displayed on the centre of a computer screen, and asked to rate the valence of the picture. The rating scale ranges from 1 (not at all disgusting) to 7 (completely

disgusting). The pictures were taken both from the International Affective Pictures System (IAPS) (Lang, Bradley, & Cuthbert, 2008) and from the web in order to have amputation images concerning both legs and arms. Four images were presented for each control emotional category, while for disgust we presented 24 images, 4 related to food, 4 to body products, 2 animal-related, 2 to contamination, 4 to death, 4 to body envelope violations (legs: 2; arms: 2) and 4 images to hygiene. Due to the difficulty to find disgusting yet legal images we did not include a category of sexually inappropriate acts. To make the analysis more meaningful, we grouped the target domains in three categories: amputation images, other domains (including images related to food, body products, animals, contamination, death and hygiene), and control images.

We presented the task on the same computer that had presented the FER task. Stimulus presentation was controlled through a custom developed Matlab© script. On each trial, one image was shown in the center of the screen (randomized order). With their dominant hand, participants had to press the keyboard number that corresponded to their disgust rating of the image, as quickly and accurately as possible. Labels were placed on the computer keyboard in a fixed order, from key "s" to key "k" of an Italian keyboard, each label depicting a number ("s" corresponding to 1). Starting point for the responding hand was always in the same position, indicated by a cross placed on the computer touchpad. Subjects were instructed to return their hand to this starting position between trials. Between trials, a fixation cross with a variable inter-stimulus interval appeared at the center of the screen. There was no time limit to complete a trial. For each trial, the rating of the image was collected. The experiment took about 10 minutes to complete.

DS

All subjects **completed** the DS (Haidt et al., 1994), a self-report questionnaire assessing disgust sensitivity in response to the verbal description of various disgust-arising situations. **The average score is approximately 16 (14 for men, 18 for women), with higher numbers indicating that a person**

is more disgust sensitive than the average population (Haidt et al., 1994). A cut off can be established at 16 for men, 20 for women (2 standard deviations above the average for the normal population).

Data analysis

Quantitative data were analysed using the program Singlims_ES (Crawford, Garthwaite, & Porter, 2010), that allows testing whether an individual's score is significantly different from a control sample. Thus, each subject with BIID has been compared to the control group by means of the Crawford t test for single subjects' comparisons (Crawford et al., 2010). See Supplementary materials (B) for a detailed discussion on this choice. Alpha level was set at 0.05. Statistical values and the estimated percentage of the normal population falling below an individual's score are reported. For the FER task, intensities have been collapsed. For both tasks, we considered the one-tailed probability value (individuals with BIID *less* accurate than controls and *diminished disgust* for emotional images related to the body).

Results

Semi-structured psychiatric interview

Most of our subjects presented with dysphoria specifically related to the amputation desire and not concerning other life domains. None of the subjects attributed their desire to a primary sexual cause. Only one individual (subject 1) showed an elevated score at the semi-structured psychiatric interview, even though his clinical history did not suggest any psychiatric disorder.

All subjects reported their amputation desire as a constant component of their life, manifesting since childhood (men reported the desire starting at age 6-7 years, the female individual at age 3-4 year).

All of them spent a variable time on the web, mostly on BIID forums, never exceeding two hours per day on average.

FER task

Subjects 1, 2, 3 and 5 did not score significantly different for overall emotion recognition (all $p > 0.05$), when compared to the control group.

Subjects 4 and 6, both males seeking for a left leg amputation, showed a similar behavioural pattern. Both were less accurate in recognizing fear (Subject 4: $t = -6.168$, estimated percentage of normal population falling below case's score (EPP): 0.008%, $p = 0.00008$; Subject 6: $t = -2.801$, EPP: 1.03%, $p = 0.01$) and disgust (Subject 4: $t = -3.400$, EPP: 0.39%, $p = 0.007$; Subject 6: $t = -2.301$, EPP: 2.34%, $p = 0.04$). Subject 6 was also less accurate for anger ($t = -2.952$, EPP: 0.80%, $p = 0.007$). Subject 7 performed less accurately than matched controls only on fear recognition ($t = -5.050$, EPP: 0.03%, $p = 0.0003$).

In summary, our results did not demonstrate a global impairment in facial emotion recognition in individuals with BIID.

PRT

First, we analyzed differences in ratings of control images, as this could be an index of test unreliability. We did not find any significant difference in these ratings (all $p > 0.05$).

When considering the “amputation” category, the female individual (subject 5) seeking for her left arm amputation, and subject 1, showing an elevated score at the semi-structured psychiatric interview and a bilateral amputation desire, did not show any significant difference in ratings when compared to control subjects (all $p < 0.05$). All the remaining individuals rated these images as less

disgusting (Subject 2: $t = -4.788$, **EPP**: 0.15%, $p = 0.0001$; Subject 3: $t = -4.172$, **EPP**: 0.29%, $p = 0.002$; Subject 4: $t = -2.326$, **EPP**: 2.94%, $p = 0.029$; Subject 6: $t = -2.326$, **EPP**: 2.94%, $p = 0.029$; Subject 7: $t = -4.172$, **EPP**: 0.29%, $p = 0.002$).

Importantly, when considering the other non-body domains (food, body products, hygiene, animal, death and contamination), we found that subject 1 rated **food images** as less disgusting when compared with controls ($t = -2.088$, **EPP**: 4.08%, $p = 0.04$). Subject 2 showed differences with respect to controls too, but in the opposite direction, rating food images and hygiene images as *more* disgusting (Food: $t = 2.262$, **EPP**: 96.78%, $p = 0.03$; Hygiene: $t = 2.211$, **EPP**: 96.54%, $p = 0.03$).

In summary, male individuals with a unilateral amputation desire considered amputation images as less disgusting (Figure 2). We emphasize that this result was found own for all four individuals singly, in contrast to the inconstant pattern found for the FER task and for the other categories of the disgust rating task. Furthermore, the female subject with BIID and the man desiring amputation of both legs, did not show diminished disgust to body violations.

[INSERT FIGURE 2]

DS

As a group, individuals with BIID obtained an average value of 11.54 (+/- 1.69) **at the DS**. Individual scores were 12.50 for subject 1, 10.50 for subject 2 and subject 5, 13.5 for subject 3, 13.00 for subject 4, and 9.25 for subject 7. None of these subjects scored outside the normal range for disgust sensitivity (**16 for men, 20 for women**). Subject 6 scored 21.5, showing a greater sensitivity to disgust than the average population. Globally, this instrument did not detect any significant pattern of generally altered disgust sensitivity in BIID individuals.

Discussion

For the first time, we explored facial emotion recognition and emotional responses to disgusting images in individuals with BIID, a condition characterized by an unmanageable desire to amputate a limb in absence of any physical disability (First & Fisher, 2012).

Our findings demonstrate that individuals with BIID do not show a global impairment in facial emotion recognition, differently from subjects affected by psychiatric diseases, who were shown to manifest a dysfunction in this ability (Kohler et al. 2011). Individuals with BIID are competent in recognizing facial displays of emotions, even when it comes to disgust, processed not only by the amygdala but also by the basal ganglia and the insula (Adolphs, 2002). While single subjects of our sample did score slightly below the control group's average score, it is the variability among the subjects with BIID that is remarkable. This variability suggests that a purely neurological modulation of facial emotion recognition in individuals with BIID is not likely to constitute a satisfactory account of the disorder. Rather, psychological or social-environmental factors should be taken into account regarding this ability.

Similarly, we did not find altered disgust sensitivity as one might expect in case of a dysfunction of the insula, the anatomical structure convincingly shown to mediate the experience of disgust (Curtis 2013). Conversely, ratings of individuals with BIID and answers to the disgust scale were generally comparable to those of gender- and age-matched controls for categories such as those depicting blood or hygiene. Importantly, a coherent pattern of diminished disgust to images of amputations was constantly shown only in male individuals with BIID seeking a unilateral amputation, while the female subject and the subject seeking a bilateral amputation rated images depicting amputations as disgusting as did controls. A possible explanation might be gender-related in the case of the female subject, and rest on the psychiatric profile in the case of the male subject. However, this explanation requires two causes for the same effect and appears thus not sufficiently parsimonious. Notably, these two individuals were the only ones showing divergent clinical features concerning the target limb to be amputated (female subject: arm, male subject: both legs). On an alternative account, our

results may suggest that diverse behaviours manifest as a function of the clinical features of the condition. For instance, individuals with unilateral BIID may be part of a unique syndrome (neurologically-based), while other individuals' condition may still require additional factors. However, whether the behavioural differences uncovered in the present examination are really related to different neural underpinnings remains to be proven.

Recent studies suggest that the majority of persons with this disorder seek for amputation as they do not perceive their body as “complete” and not based on a sexual impulse (First, 2012). Notably, this neurological explanation for BIID is not uniformly supported in the scientific community. An alternative hypothesis to the neurological one suggests a psychiatric origin of BIID (De Preester, 2011). Patients with psychiatric disturbances frequently experience an altered sense of bodily self (Ferri et al., 2012), framed into more generalized emotional disturbances. For example, both bipolar syndrome and depression are featured by moderate but constant deficits in facial emotion recognition (Kohler et al., 2011).

If BIID represents a psychiatric condition, one might expect a pervasive emotional impairment: less accurate facial emotion recognition and no disgust for images that normally elicit this emotion. For instance, it has been suggested that emotional deficits in schizophrenia might be due to a “polymodal disruption in sensory-affect processing” attributable to an impaired insular functionality (Wylie & Tregellas, 2010). Our results do not support this aetiology in spite of the reported insular involvement in BIID (Hilti et al., 2013): findings from the FER task and from the **PRT** do not support a global impairment in emotion recognition and in emotional responding in these individuals.

One might hypothesize that these more complex emotional aspects may be spared in persons with BIID, given that their attention is focused on a body part, suggesting a disorder selectively confined to the emotional components of the body representation. In this sense, BIID would be different from other psychiatric disorders that are characterized by widespread deficits rather than by specific

impairments (Kohler et al., 2011). The selective **diminished disgust** for amputation images seems to point in this direction, corroborating the idea that BIID is a specific disorder confined to the bodily self. **Body image refers to the "system of perceptions, attitudes, and beliefs pertaining to one's own body" (Gallagher, 2005, p.24),** and its processing is suggested to be modulated by the insular cortex (Berlucchi & Aglioti, 2010). We demonstrate that violations of the ideal body are not experienced as disgusting by persons with BIID (unilateral amputation desire), and a possible dysfunction in the insular region has been associated with this condition. Consequently, the insular involvement in BIID might be more related to the internal state processing rather than to a problem in polymodal sensory-affect processing. The insular cortex modulates interoception and seems to be involved in high order re-representations of the internal state that contributes to our body representation (Craig, 2009). **Firstly, an emotional feeling is generated through a neurovisceral response to a certain stimulus and is then evaluated as being of a positive/negative valence (Lang et al. 1998). Sensitivity of these physiological responses is mediated by interoceptive awareness, and might be one of the neural mechanisms underlying the reduced response to disgusting images. The reduction in disgust sensitivity specific to body-violations points to the fact that it might be a higher order alteration in emotional appraisal rather than a general neurovisceral reaction. This hypothesis should be examined in future studies with measurement of autonomic responses to such stimuli (for instance, galvanic skin responses).**

Further, the anterior insula is known to be associated with worry processing (Paulus & Stein, 2006) and is part of a widespread circuit also involving prefrontal and limbic structures and the ventral striatum, considered as a "compulsivity circuit" (Naqvi, Rudrauf, Damasio, & Bechara, 2007). Not by chance, greater activations of the right insula compared to normal controls have been found in obsessive compulsive disorder (OCD) patients when watching disgusting pictures (Shapira et al., 2003). BIID as well is characterized by recurrent thinking and worry, although featuring a mirrored pattern when compared with OCD: individuals with BIID, distressed by the overwhelming desire of

amputation, seek for surgery even though facing risky and unsafe situations or even undergo self-amputations. Conversely, **OCD** patients typically develop an excessive preoccupation for violations of the hygienic norms and apply increasingly obsessive behaviours of disease avoidance.

The association of our PRT and of instruments to assess worry and recurrent thinking with a functional MRI technique could provide a less speculative evidence of an insular involvement in BIID as far as emotional components of body representation are concerned. Whenever this postulated association between insular functioning and BIID will be made more plausible, it remains an open question whether this dysfunction represents the cause or the consequence of BIID. An alternative interpretation of our results could suggest an inverse relationship with empathy. Many persons with BIID describe being drawn to other amputees, either through admiration (e.g., for overcoming adversity) or through sexual ideation (Bou Khalil & Richa, 2012). This might relate to increased empathy and/or sensorimotor resonance for individuals with an amputation and, consequently, **with a diminished disgust response to amputations.** **Empathy includes somatic and visceral responses together with cognitive components (Preston & de Waal 2002), and when empathizing positively with a stimulus or a person, the negative reaction decreases.** Thus, it could also be that BIID modifies insular functioning during the lifetime, **substituting disgust with empathic responses for selective categories of normally disgust-provoking stimuli.** Surely this interpretation is intriguing and suggests a more scrutinizing analysis of higher emotional abilities in persons with BIID.

One might object that the overexposure to amputation images, when navigating the web, or the reiterative thinking related to the amputation desire since its onset can explain the reduced response to the body envelope, as if individuals with BIID developed an habituation effect. **However, our subjects reported to spend no more than 2 hours a day on the web (see semi structured interview), a time that can be hardly considered an overexposure.** Similarly, the time of the onset of the amputation desire was comparable across subjects; all of them referred the origin of their desire to

childhood. Moreover, disgust is known to be quite resistant to extinction and habituation, even when explicit self-report measures were employed (Broderick, Grisham, & Weidemann, 2013).

Although our behavioural evidence is preliminary, we do think that it allows questioning a purely neurological origin of BIID. Likewise, our results do not support a pure psychiatric explanation either, at least not one grounded on the classical clinical taxonomy. Conversely, our results indicate that BIID represents a clinical constellation. The heterogeneity of individuals with BIID is becoming more and more apparent. Among those desiring amputation, only some indicate that they would use prostheses after the operation. Among those who seek for paraplegia, some desire bladder paralysis as well, while others would abhor any dysfunction apart from limb paralysis. On top of all these differences, gender seems to be a further modulating factor (Giummarra et al. 2012).

Our results strongly agree with this last finding, as the female individual presented a different rating of amputation images. Even though most studies have hitherto considered BIID a unique entity (see for instance McGeoch et al., 2011), we adopted a multiple single cases approach strategy. This choice proved efficient, allowing us to identify a pattern of results coherent for subjects with comparable clinical features. Nevertheless, one limitation of our study is the number of individuals we have been able to test. Such a small and heterogeneous sample only allows the performance of single case comparisons. Future studies should recruit more subjects, grouped into subgroups representing comparable clinical categories (i.e. individuals who seek a unilateral amputation of the leg versus individuals who seek a unilateral amputation of the arm). Further, knowledge about BIID would greatly profit from the inclusion of variants, in which subjects do not seek amputation of the limb but rather paraplegia (Giummarra et al. 2012). One might reason that this is a different form of body violation, and thus it might induce different responses to disgust related images as well. In these individuals, viewing amputation images may elicit comparable responses to control samples, as people who desire paraplegia would typically not accept amputation as an alternative.

Considering the paradigms that could be employed when studying emotional processing in individuals with BIID, our study did not include measures of high emotional competences such as empathy. Nevertheless, our findings could well allow inferences about empathy and the feeling of being **attracted to amputees**. Thus, future work could consider including more tasks, tackling theory of mind and other social competences. Further, the possible modulation of the social factors of the environment should be taken into account as well, being a component of a complex model of body representation that comprises emotions, not only in their individual basic aspects but also in a social-biological framework (Brugger, Lenggenhager, & Giummarra, 2013).

A final remark concerns the level of measurement we employed. Our tasks required conscious responses to emotional stimuli. However, it might be of interest to explore whether conscious and unconscious components of emotional processing dissociate in individuals with BIID. The contribution of the two hemispheres to emotional processing has been suggested to be different depending on the level of processing. The right hemisphere regulates unconscious aspects, while conscious aspects, more available to verbal awareness, are rather mediated by the left hemisphere (Gainotti, 2012). Adopting tasks exploring both levels would shed further light on brain lateralization in BIID, as available studies suggest a major involvement of the right hemisphere. Given the undeniable fact that BIID symptoms target the left side of the body more than the right, one would expect that especially right hemisphere mediated, implicit components of emotional processing should be compromised.

In conclusion, our study highlights the importance to consider aspects of body representation that are not properly spatial-corporeal, but overlap with morality and social cognition. Even though preliminary, our data are the first available on emotion recognition and disgust processing in BIID. As such they provide a number of possible future directions that could contribute to the understanding of the aetiology of this condition. Further, we demonstrate that when taking into account emotion-related aspects, stable differences emerge between individuals, in congruency with

their diverse clinical features and their gender. Thus, caution is warranted when studying BIID, as this condition appears more and more a complex syndrome comprising cognition and emotion and their complex interactions.

References

- Adolphs, R. (2002). Neural systems for recognizing emotion. *Curr Opin Neurobiol*, 12(2), 169-177.
- Berlucchi, G., & Aglioti, S. M. (2010). The body in the brain revisited. *Exp Brain Res*, 200(1), 25-35.
- Bottini, G., Sedda, A., Ferre, E. R., Invernizzi, P., Gandola, M., & Paulesu, E. (2009). Productive symptoms in right brain damage. *Curr Opin Neurol*, 22(6), 589-593.
- Bou Khalil, R., & Richa, S. (2012). Apotemnophilia or body integrity identity disorder: a case report review. *Int J Low Extrem Wounds*, 11(4), 313-319.
- Brang, D., McGeoch, P. D., & Ramachandran, V. S. (2008). Apotemnophilia: a neurological disorder. *Neuroreport*, 19(13), 1305-1306.
- Broderick, J., Grisham, J. R., & Weidemann, G. (2013). Disgust and fear responding in contamination-based obsessive-compulsive disorder during pictorial exposure. *Behav Ther*, 44(1), 27-38.
- Brugger, P., Lenggenhager, B., & Giummarra, M. J. (2013). Xenomelia: a social neuroscience view of altered bodily self-consciousness. *Front Psychol*, 4, 204.
- Craig, A. D. (2009). How do you feel -- now? The anterior insula and human awareness. *Nat Rev Neurosci*, 10(1), 59-70.
- Crawford, J. R., Garthwaite, P. H., & Porter, S. (2010). Point and interval estimates of effect sizes for the case-controls design in neuropsychology: rationale, methods, implementations, and proposed reporting standards. *Cogn Neuropsychol*, 27(3), 245-260.

- Curtis, V. (2011). Why disgust matters. *Philos Trans R Soc Lond B Biol Sci*, 366(1583), 3478-3490.
- De Preester, H. (2011). Merleau-Ponty's sexual schema and the sexual component of body integrity identity disorder. *Med Health Care Philos*, 16(2):171-84.
- Ekman, P., & Friesen, W. V. (1976). *Pictures of Facial Affect*. Palo Alto: Consulting Psychologist Press.
- Ferri, F., Frassinetti, F., Mastrangelo, F., Salone, A., Ferro, F. M., & Gallese, V. (2012). Bodily self and schizophrenia: the loss of implicit self-body knowledge. *Conscious Cogn*, 21(3), 1365-1374.
- First, M., Gibbon, M., Spitzer, R., Williams, J., & Benjamin, L. (1997). *Structured Clinical Interview for DSM-IV Axis II Personality Disorders (SCID-II)*. Washington, D.C.
- First, M. B., & Fisher, C. E. (2012). Body integrity identity disorder: the persistent desire to acquire a physical disability. *Psychopathology*, 45(1), 3-14.
- Gainotti, G. (2012). Unconscious processing of emotions and the right hemisphere. *Neuropsychologia*, 50, 205–218.
- Gallagher, S. (2005). *How the Body Shapes the Mind*. New York: Oxford University Press.
- Gandola M., Invernizzi P., Sedda A., Ferrè E.R., Sterzi R., Sberna M., Paulesu E., & Bottini G. (2012). An anatomical account of somatoparaphrenia. *Cortex*, 48(9), 1165-1178.
- Giummarra MJ, Bradshaw JL, Hilti LM, Nicholls ME, & Brugger P. (2012). Paralyzed by desire: a new type of body integrity identity disorder. *Cogn Behav Neurol*, 25(1), 34-41.
- Giummarra, M. J., Bradshaw, J. L., Nicholls, M. E., Hilti, L. M., & Brugger, P. (2011). Body integrity identity disorder: deranged body processing, right fronto-parietal dysfunction, and phenomenological experience of body incongruity. *Neuropsychol Rev*, 21(4), 320-333.
- Haidt, J., McCauley, C., & Rozin, P. (1994). Individual differences in sensitivity to disgust: A scale sampling seven domains of disgust elicitors. *Personality and Individual Differences*, 16, 701-713.

- Hilti L.M., Hänggi J., Vitacco D.A., Kraemer B., Palla A., Luechinger R., Jäncke L., & Brugger P. (2013). The desire for healthy limb amputation: structural brain correlates and clinical features of xenomelia. *Brain*, 136(Pt 1), 318-329.
- Jabbi, M., Bastiaansen, J., & Keysers, C. (2008). A common anterior insula representation of disgust observation, experience and imagination shows divergent functional connectivity pathways. *PLoS One*, 3(8), e2939.
- Kohler, C. G., Hoffman, L. J., Eastman, L. B., Healey, K., & Moberg, P. J. (2011). Facial emotion perception in depression and bipolar disorder: a quantitative review. *Psychiatry Res*, 188(3), 303-309.
- Lang, P. J., Bradley, M. M., & Cuthbert, B. N. (2008). *International affective picture system (IAPS): Affective ratings of pictures and instruction manual*. Technical Report A-8. Gainesville, FL.
- Lang PJ, Bradley MM, & Cuthbert B.N. (1998). Emotion, motivation, and anxiety: brain mechanisms and psychophysiology. *Biol Psychiatry*, 44(12):1248-63.
- Lenggenhager B., Hilti L., Palla A., Macaudo G. & Brugger O. (2014). Vestibular stimulation does not diminish the desire for amputation. *Cortex* (in press).
- McGeoch, P. D., Brang, D., Song, T., Lee, R. R., Huang, M., & Ramachandran, V. S. (2011). Xenomelia: a new right parietal lobe syndrome. *J Neurol Neurosurg Psychiatry*, 82(12):1314-9.
- Naqvi, N. H., Rudrauf, D., Damasio, H., & Bechara, A. (2007). Damage to the insula disrupts addiction to cigarette smoking. *Science*, 315(5811), 531-534.
- Paulus, M. P., & Stein, M. B. (2006). An insular view of anxiety. *Biol Psychiatry*, 60(4), 383-387.
- Phillips K.A., Menard W., Fay C., & Weisberg R. (2005). Demographic characteristics, phenomenology, comorbidity, and family history in 200 individuals with body dysmorphic disorder. *Psychosomatics*, 46(4), 317-25.

Preston S.D. & de Waal F.B. (2002). Empathy: its ultimate and proximate bases. *Behav. Brain Sci.*, 25,1–20.

Rapcsak S.Z., Galper S.R., Comer J.F., Reminger S.L., Nielsen L., Kaszniak A.W., Verfaellie M., Laguna J.F., Labiner D.M., & Cohen R.A. (2000). Fear recognition deficits after focal brain damage: a cautionary note. *Neurology*, 54(3), 575-581.

Rozin, P., Haidt, J., & Fincher, K. (2009). Psychology. From oral to moral. *Science*, 323(5918), 1179-1180.

Ryan C.J. & Shaw T. (2011). BIID is not BDD--comment on Kwok-Kwan Chan et al. *J Plast Reconstr Aesthet Surg*, 64(1), 8-9.

Schienze, A., Haas-Krammer, A., Schoggl, H., Kapfhammer, H. P., & Ille, R. (2013). Altered state and trait disgust in borderline personality disorder. *J Nerv Ment Dis*, 201(2), 105-108.

Sedda, A. (2011). Body integrity identity disorder: from a psychological to a neurological syndrome. *Neuropsychol Rev*, 21(4), 334-336.

Sedda, A., Rivolta, D., Scarpa, P., Burt, M. D., Frigerio, E., Zanardi, G., Piazzini A., Turner K., Canevini M.P., Francione S., Lorusso G., & Bottini, G. (2013). Ambiguous emotions recognition in temporal lobe epilepsy: the role of expression intensity. *Cogn Affect Behav Neurosci*, 13(3):452-63.

Shapira N.A., Liu Y., He A.G., Bradley M.M., Lessig M.C., James G.A., Stein D.J., Lang P.J., & Goodman W.K. (2003). Brain activation by disgust-inducing pictures in obsessive-compulsive disorder. *Biol Psychiatry*, 54(7), 751-756.

Figure captions

Figure 2. Ratings at the PRT. Data are presented separated for males and females. For the control group, average rating (and standard deviation) is depicted. Stars indicate a significantly different rating compared with the control group. The third graph present data from individuals with BIID as a group. Subj = subject.

